Figure 1

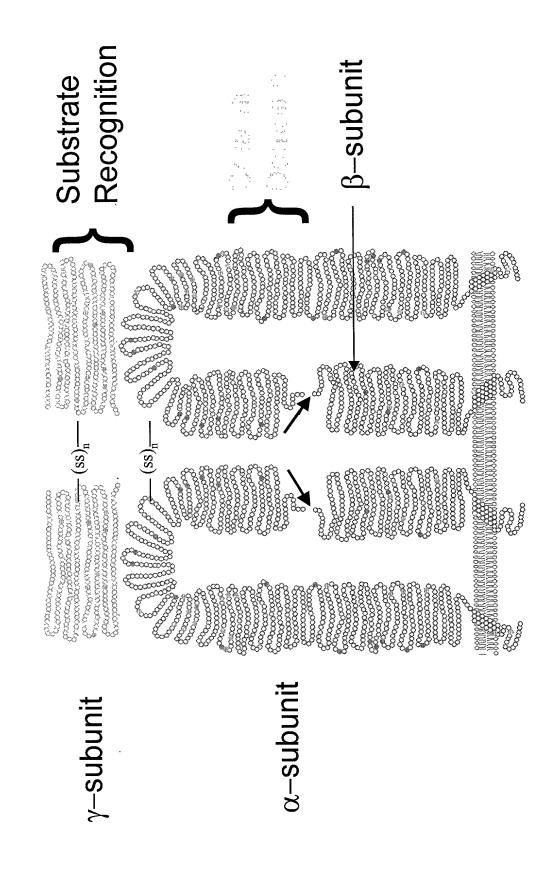
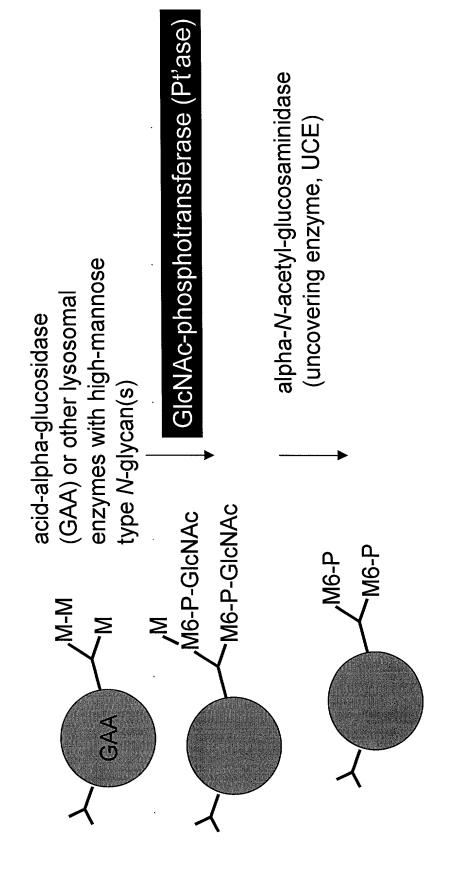


Figure 2



After phosphorylation/uncovering, lysosomal enzyme is recognized by mannose-6-phosphate receptor (M6PR) and targeted to lysosome.



GlcNAc-phosphotransferase

wild type  $\alpha/\beta$  subunit

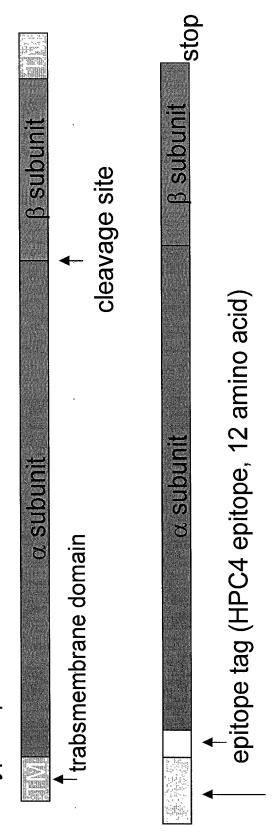


Figure 4

## Figure 5

------DEEESLKTQLAYFTDSKNTGRQLKDTFADSLRYVNKILNSKFGFTS--Factor Xa beta subunit alpha subunit wild type

---- FTDSKNTG IEGRDTFADS----

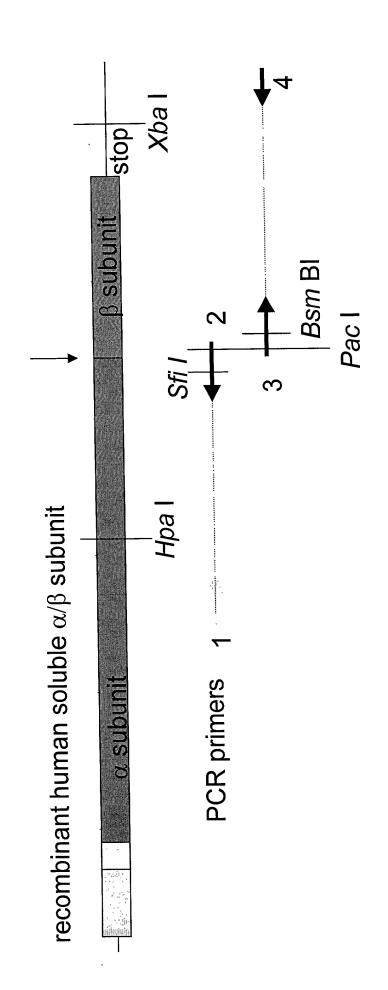
Enterokinase ---- FTDSKNTDDDDKDTFADS----

---- FTDSKNRARYKRDTFADS----

Furin

Genease ---- FTDSKNPGAAHYDTFADS----

Figure 6A



## Figure 6B

5'-GAGGCAATTAATTAACGTCTCTGCAGATTCCCTCAGATATGTAAATAA-3'

S'-GAGGCAATTAATTAACGTCTCTGCAGATTCCCTCAGATATGTAAATAA-3' 3'-CTCAGTAACTTCTGTGTTAACCGGATGAACCGG<u>AATTAATT</u>CTTATGAC-5'

Pac | BsmB

PCR primer 2

PCR primer 3

EEESLKTQLA

ADLSLRYVNKI

Pac I BsmB Sfil

## Figure 6C



Enterokinase Furin ---ACACAATTGGCCTACTTCACTGATAGCAAGAATAGAGCCAGATACAAGAGAGATACATTTGCAGATTCCCTCAGA---T Q L A Y F T D S K N T D D D D K D T F A D S L R ADSLR T Q L A Y F T D S K N R A R Y K R D T F A D S L R QLAYFTDSKNPGAAHYDTF  $\vdash$ 

SLR TQLAYFTDSKNTGRQLKDTFAD

Wild typee Genease

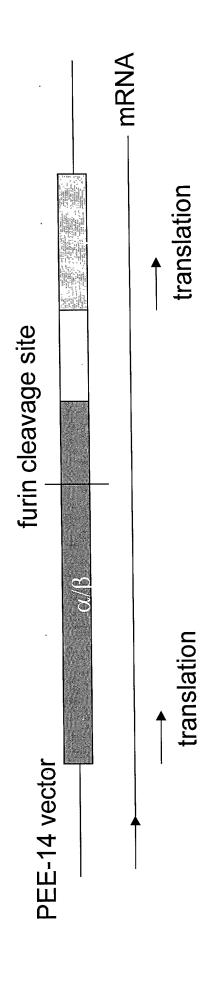


Figure 7